Imaging and clinical features of uncommon primary mediastinal masses

Poster No.: C-1197
Congress: ECR 2015
Type: Educational Exhibit
Authors: Y. Sato¹, K. Nakanishi¹, N. Tomiyama²; ¹Osaka/JP, ²Sutta/JP
Keywords: Education, MR, CT, Mediastinum, Cancer
DOI: 10.1594/ecr2015/C-1197

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Learning objectives

1. List the various uncommon primary mediastinal masses.

2. Recognize the clinical and imaging characteristics of these entities.

3. Discuss some of the imaging features that help the radiologist to narrow the differential diagnosis of mediastinum.

Background

There is a wide spectrum of mediastinal tumor which includes, thymoma, malignant lymphoma, neurogenic tumor, teratoma, bronchogenic cyst, etc.

Rarely mediastinal sarcomas, neuroendocrine tumors, germinal cell tumors and uncommon cystic masses are encountered. Some of these clinical and imaging characteristics may facilitate narrowing differential diagnosis.

Findings and procedure details

In this presentation, we review our experience with rare mediastinal masses.

We also correlate the imaging features with the clinical and pathologic features.

The spectrum of rare mediastinal masses according to different pathological entities is summarized below.

1. Neuroendocrine tumors of the thymus

Carcinoid

Large cell neuroendocrine carcinoma (LCNEC)

2. Primary mediastinal sarcomas
Liposarcoma
Ewing sarcoma
Malignant fibrous histiocytoma (MFH)
Fibromyxoid sarcoma
Synovial sarcoma
3. Primary mediastinal germ cell tumors
Immature teratoma
Seminoma
Choriocarcinoma
4. Cyst-like lesions of the mediastinum
Thoracic lateral meningocele
Lymphatic malformation

1. Neuroendocrine tumors of the thymus

Neuroendocrine tumors of the thymus are categorized within the group of thymic carcinomas.

Although thymic carcinomas account for approximately 20% of thymic epithelial tumors, almost all thymic carcinomas are squamous cell carcinoma pathologically, and thymic neuroendocrine tumors are rare\(^1\). Thymic neuroendocrine tumors are classified into four groups: typical carcinoid, atypical carcinoid, small cell carcinoma, large cell neuroendocrine carcinoma (LCNEC).

#Carcinoid of the thymus

Thymic carcinoid predominantly occurs in the anterior mediastinum.

It is found more often in the fourth and fifth decades of life.

Approximately 50% of patients have endocrine disorders, such as Cushing syndrome (20-40% of patients) or multiple endocrine neoplasia syndrome (19-25% of patients). It is reported that thymic carcinoid with multiple endocrine neoplasia syndrome (MEN) type 1 develops in 8% of cases, and 95% of patients are male, compared to the rate of 40% of MEN type 1 lesions without a thymic carcinoid component\(^2,3\).
Most thymic carcinoids exhibit low-grade malignancy and display a tendency toward invasion, local recurrence after resection and/or metastasis to regional lymph nodes or distant organs\(^4\).

**Imaging findings** (Fig. 1)

Thymic carcinoid manifests as large, lobulated, invasive masses that may show areas of low attenuation due to hemorrhage and necrosis\(^3\). The appearance of these lesions on CT scans is nonspecific, and it is difficult to distinguish between thymic carcinoids and other mediastinal masses, such as thymomas, germ cell tumors and malignant lymphomas.

**Key points**

If a patient with MEN type 1 or Cushing syndrome has an anterior mediastinal mass, a carcinoid may be suspected.

**#Large cell neuroendocrine carcinoma (LCNEC) of the thymus**

LCNECs are more highly malignant and have a poorer prognosis than carcinoids. Patients with LCNEC frequently develop recurrence within a short period of time and display a high rate of distant relapse\(^5\).

**Imaging findings** (Fig. 2)

There have been no reviews regarding the imaging findings of LCNEC of the thymus. A few case reports are seen, and show non-specific anterior mass.

**2. Primary mediastinal sarcoma**

Primary sarcomas of the mediastinum are rare.

Until the tumors reach a remarkable size, they do not produce symptoms. Imaging findings have been reported in only few cases.

**#Liposarcoma**

Liposarcomas are reported to represent 0.1-0.75% of all primary mediastinal tumors. Liposarcomas appear most commonly in patients over 40 years of age. Almost all liposarcomas occur in the posterior or anterior mediastinum and usually present with symptoms at the time of diagnosis, in contrast to lipomas.

**Imaging findings** (Fig. 3)
Liposarcomas demonstrate higher CT attenuation values than lipomas (from -30 to -100 HU). When the CT attenuation value exceeds -30 HU, a diagnosis of liposarcoma may be suspected\(^6,7\). An inhomogeneous appearance on CT or MRI discriminates liposarcomas between lipomas\(^8\). Liposarcomas exhibit a wide range of differentiation, from low to high grade. Lower CT values for liposarcomas correlate with better differentiation, while more poorly differentiated liposarcomas demonstrate well-enhanced soft tissue attenuation induced by contrast media\(^1\).

Fat-containing tumors of the mediastinum are limited, with teratomas being the most frequent.

Fifty-three percent of patients with mature teratomas have been reported to have characteristic sites of calcification, a finding that may be helpful for distinguishing these lesions from liposarcomas\(^9\). Fat components may not be detected in liposarcomas, which makes difficult to discriminate liposarcomas from other mediastinal masses.

**Key points**

Liposarcomas are fat-containing tumors with an inhomogeneous appearance on CT or MRI and well-enhanced soft tissue attenuation induced by contrast media.

Without the presence of fat components, it is difficult to discriminate liposarcomas from other mediastinal masses.

### Ewing sarcoma

The prevalence of extraskeletal Ewing sarcoma is generally accepted to be between 15% and 20% of that of bone Ewing sarcoma\(^10\).

Although Ewing sarcoma usually presents in the second decade of life, one study of Ewing sarcoma arising in the mediastinum reported that the average age at diagnosis was 40 years\(^11\). Mediastinal Ewing sarcomas may therefore tend to occur in adults.

**Imaging findings** (Fig. 4)

Ewing sarcomas present as ill-defined, heterogeneous masses. These findings are nonspecific, and the lesions cannot be distinguished from other mediastinal tumors.

**Key points**

For children and young adults presenting with multifocal intrathoracic mass lesions, extraskeletal Ewing sarcoma may be considered in the differential diagnosis. However, mediastinal Ewing sarcomas may be inclined to occur in adults, and it may be difficult to distinguish these lesions from the other mediastinal tumors.
# Malignant fibrous histiocytoma (MFH)

Although MFH is the most common malignant soft tissue sarcoma in adults, MFH of mediastinal origin is extremely rare.

Most cases of MFH occur in older adults. The prognosis is poor, with a high incidence of local recurrence and distant metastasis.

**Imaging findings** (Fig. 5)

MFH usually involves well-marginated, smooth or lobular, soft tissue masses. MFH lesions show heterogeneous attenuation and enhancement on CT\(^{12}\). Calcification is uncommon prior to therapy.

**Key points**

Although MFH exhibits nonspecific findings, MFH lesions are difficult to discriminate from other tumors of the mediastinum.

# Fibromyxoid sarcoma

Fibromyxoid sarcoma is usually found in young and middle-aged adults in the form of large masses characterized by fibrous, myxoid areas and may be located in the mediastinum. These tumors are usually slow growing and can recur or metastasize over a long period after resection.

**Imaging findings** (Fig. 6)

Fibromyxoid sarcomas have large well-defined margins with heterogeneous attenuation and enhancement on CT. On T1- and T2-weighted MR images, the masses display heterogeneous intensity and enhancement correlated with myxoid and fibrous portions. The tumors have a multinodular structure and components with an abrupt transition, indicating that the lesion may be a fibromyxoid sarcoma\(^{13}\).

**Key points**

The presence of a multinodular structure and components with an abrupt transition in mediastinal tumors suggests that the lesion may be a fibromyxoid sarcoma.

# Synovial sarcoma

Primary mediastinal mesenchymal tumors are reported to represent 0.7% of all primary mediastinal tumors; synovial sarcomas in particular are extremely rare.
These lesions occur in young adults (20 to 30 years of age).

**Imaging findings** (Fig. 7)

Synovial sarcomas typically manifest as heterogeneous masses that occasionally contain calcium. On MR imaging, these tumors demonstrate heterogeneous signal intensity on T1- and T2-weighted images and may have fluid-fluid levels due to the presence of hemorrhage and necrosis within cystic components of the tumor\(^{12}\).

**Key points**

Although synovial sarcomas can be difficult to distinguish from other soft tissue tumors, calcification is seen in approximately 30% of lesions, and the detection of heterogeneous signal intensity on T1- and T2-weighted images may be helpful for differentiating these tumors.

3. **Primary mediastinal germ cell tumors**

Germ cell tumors (GCTs) mainly arise in the gonads and the mediastinum being the most common extragonadal site. Primary mediastinal germ cell tumors frequently occur in the anterior mediastinum. Pathologically they are classified into teratomas and malignant germ cell tumors, the latter of which consists of seminomas, yolk sac tumors, embryonal carcinomas, choriocarcinomas and mixed germ cell tumors.

More than half of mediastinal germ cell tumors are mature teratomas.

**#Immature teratoma**

Teratoma is the most common subtype of GCT. Although almost teratomas are mature teratomas that show a benign course, teratomas rarely contain fetal tissue, classified as immature teratomas, which have a good prognosis in children, although they may recur or metastasize.

**Imaging findings** (Fig. 8)

Teratomas have been reported to contain fat (76% of patients), fluid (88%) and calcification (53%), and their specific components may help to differentiate these lesions from other mediastinal masses\(^9\). Without these components, it is difficult to distinguish teratomas from other masses.

The presence of solid tissue and a thick capsule within the lesion is more commonly seen in immature malignant teratomas than in mature benign tumors. Mature benign teratomas tend to displace adjacent structures, whereas malignant immature teratomas tend to invade these tissues\(^{14}\).
Key points

Immature teratomas tend to more frequently contain solid tissue and a thick capsule within the lesion and to be more invasive than mature benign teratomas.

*Malignant germ cell tumors*

Among malignant mediastinal germ cell tumors, 40% are seminomas and 60% are non-seminomatous tumors. Yolk sac tumors are the most common mediastinal non-seminomatous tumor (60%), followed by mixed tumors (18%), choriocarcinomas (12%) and embryonal carcinomas (9%).

Recent reports have revealed that mediastinal malignant germ cell tumors usually occur in young males. Patients with such tumors exhibit no particular clinical manifestations, which results in the detection of bulky masses upon referral to the hospital for treatment.

**Imaging findings**

Seminomas are well-circumscribed masses with a lobulated shape as well as homogeneous attenuation on non-enhanced CT images and homogeneous enhancement after contrast administration (Fig. 9). In contrast, non-seminomatous tumors are bulky, ill-circumscribed, lobulated anterior mediastinal masses displaying areas of heterogeneous attenuation with central necrosis, hemorrhage and/or cyst formation and calcification on non-enhanced CT images in addition to heterogeneous enhancement after the administration of contrast material (Fig. 10). Although these findings are also observed in other mediastinal masses, such as thymomas, thymic carcinomas and lymphomas, it is difficult to distinguish between these tumors. Nevertheless, elevation of serum tumor markers, i.e., #-fetoprotein or #-fraction of human chorionic gonadotropin, is suggestive of malignant germ cell tumors, whereas an increased AFP level indicates an embryonal carcinoma or Yolk sac tumor, a very high hCG level reflects choriocarcinoma and a mildly elevated hCG level is indicative of seminoma (10% of patients).

Key points

#Seminomas show homogenous attenuation and enhancement following contrast administration.

#Non-seminomatous tumors exhibit inhomogeneous attenuation and enhancement after contrast administration.

#An elevated hCG level indicates choriocarcinoma and an elevated level of AFP suggests embryonal carcinoma or a Yolk sac tumor.
4. Cyst-like lesions of the mediastinum

Mediastinal cyst-like masses are well-defined round lesions containing fluid and lined with epithelium. Major cystic masses include congenital benign cysts (e.g. bronchogenic, pericardial and thymic cysts, etc.). Uncommon mediastinal cystic masses (e.g. meningocele, lymphatic malformation and so on) are seen in the mediastinum.

#Lateral thoracic meningocele

Lateral thoracic meningocele is a rare posterior mediastinal cystic lesion, defined as the saccular protrusion of the spinal meninges through the intravertebral foramen. This abnormality is frequently associated with neurofibromatosis and it often occurs in the fourth to fifth decades of life.

**Imaging findings** (Fig. 11)

On CT, lateral thoracic meningocele presents as a well-defined, homogeneous, low-attenuation paravertebral mass frequently associated with osseous abnormalities, such as pressure erosion of the posterior vertebral body, widening of the neural foramina and kyphoscoliosis. Continuity between meningocele and the dural sac is seen on CT and MRI.

**Key points**

Continuity between paravertebral masses and the dural sac indicates lateral thoracic meningocele, which may be associated with osseous abnormalities.

#Lymphatic malformation

Lymphatic malformation, also known as lymphangioma, is a rare, benign congenital malformations that represents 0.7-4.5% of all mediastinal tumors. Lymphatic malformation involve the neck or the axillary region in more than 80% of the cases and the thorax in 1-10% of the cases. Most lymphatic malformations present early in life, however, they are discovered in adulthood accidentally without symptoms. Pathologically, lymphatic vessels and venous vessels may be mixed in the mass.

**Imaging findings** (Fig. 12)

Lymphatic malformations were lobulated, well-capsulated, smoothly margined and cystic, but they may have higher attenuation due to previous hemorrhage or infection producing protein on CT. The lesions have heterogeneous signal intensity on T1-weighted images, and they usually have high signal intensity on T2-weighted images reflects their fluid content. Thin and meandering septum within the mass may sometimes
be discovered. Enhancement of internal septa and the cyst wall may be seen in a mass but enhancement of central portions are not seen.

**Key points**

Lymphatic malformation usually appears multilocular cystic mass.

**Images for this section:**

![Image](image-url)

**Fig. 1:** Carcinoid in a 57-year-old male. A contrast-enhanced CT scan shows a well-defined mass on the left side of the anterior mediastinum. The patient developed hypercalcemia due to parathyroid adenoma. This indicates that he may have suffered from MEN type1.
**Fig. 2:** LCNEC in a 44-year-old male. An unenhanced CT scan (above) scan shows mass with irregular margin on the right side of the anterior mediastinum. T2-weighted imaging (below) shows a heterogeneous mass.
Fig. 3: Liposarcoma in a 69-year-old male. A contrast-enhanced CT scan (above) shows a large well-defined mass with well-enhanced soft tissue. T1-weighted imaging (below) shows a high-intensity signal due to the presence of fat and an iso-intense signal due to the presence of soft tissue.

![Image of CT scan showing liposarcoma](image)

Fig. 4: Ewing sarcoma in a 6-year-old male. An axial contrast-enhanced CT scan shows a large well-defined mass with homogeneous enhancement in the posterior mediastinum.

![Image of CT scan showing Ewing sarcoma](image)
**Fig. 5:** MFH in a 62-year-old male. A contrast-enhanced CT scan (above) shows a large well-defined mass on the right side of the anterior mediastinum with heterogeneous enhancement. T2-weighted image (below) shows an inhomogeneous mass.
**Fig. 6:** Fibromyxoid sarcoma in a 51-year-old female. A contrast-enhanced CT scan (above) shows a well-defined mass in the paravertebral region with heterogeneous enhancement. T2-weighted MR imaging (below) demonstrates an inhomogeneous mass.
**Fig. 7:** Synovial sarcoma in a 66-year-old female. An unenhanced CT scan shows a well-defined mass in the anterior mediastinum with calcification (arrow). A contrast-enhanced CT scan shows a large well-defined mass on the left side of the anterior mediastinum with heterogeneous enhancement.

**Fig. 8:** Immature teratoma in a 17-year-old female. A contrast-enhanced CT scan shows a large mass in the anterior mediastinum with nodules exhibiting heterogeneous enhancement and fat attenuation.
**Fig. 9:** Seminoma in a 37-year-old male. A contrast-enhanced CT scan shows homogeneous enhancement in the anterior mediastinum.
Fig. 10: Choriocarcinoma in a 24-year-old male. A contrast-enhanced CT scan shows mass with irregular margin and heterogeneous enhancement in the anterior mediastinum. Multiple pulmonary metastases in the bilateral lungs and pleural effusion are shown. The levels of serum tumor markers were elevated (AFP 57ng/ml, HCG 215000mIU/ml, #-hCG 1900ng/ml).
**Fig. 11:** Lateral thoracic meningocele in a 72-year-old female. The mass continued to the dural sac. The patient was complicated with neurofibromatosis.
Fig. 12: Lymphatic venous malformation in a 37-year-old female. A contrast-enhanced CT scan (above) shows a well-defined mass on the left side of the anterior mediastinum. T2-weighted image (below) show a high-intensity mass with a low-intensity portion. Pathologically, many vessels combined with the lymphatic and venous vasculature in the tumor are enlarged, as in a cyst.
Conclusion

1. Although it is difficult to determine each type of thymic cancer radiologically, the clinical characteristics may be helpful for diagnosing thymic cancer.

2. Although it is difficult to differentiate each type of sarcoma radiologically, the presence of fat attenuation may indicate liposarcoma.

3. Becoming familiar with the CT and MR imaging features of various mediastinal masses is useful for obtaining an accurate diagnosis.

Personal information

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